

## A Case of Angry Birds – Aggressive Angiomyxoma.

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### ABSTRACT

Aggressive angiomyxoma is a non metastasising soft tissue tumor of pelvis and perineum occurring in women of reproductive age group which carried a high risk for local relapse, hence the need to differentiate it from other mesenchymal tumors. We describe a case of 45 years old female presenting with large mass coming out of vagina.

**Keywords:** Aggressive angiomyxoma, mesenchymal tumor, cervix tumor.

### INTRODUCTION

Aggressive angiomyxoma is a slow growing pelvic and perineum mesenchymal neoplasm with marked tendency of local recurrence but low tendency to metastasize. It was first described by Steper and Rosai in 1983.<sup>[1]</sup> Estrogen receptors and progesterone receptors are usually found in aggressive angiomyxoma.<sup>[2]</sup> It involves mainly the pelvic, perineum, vagina and urinary bladder in adult women in the reproductive age group. Considering its locally aggressive nature, appropriate management and long term followup is necessary.<sup>[3]</sup>

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spindle cells with ovoid nuclei and ill-defined cytoplasmic border. Cellular atypia is not seen. Many thick and thin walled blood vessels are seen (Figure 3,4).



Fig:1

Figure 1: Photograph showing a cervical mass.

### CASE REPORT

A 45 year old female presented with some mass coming out of vagina along with yellowish vaginal discharge. Her menstrual history was normal and vitals were stable. Uterus was not palpable. Local examination revealed a mass which seems to be arising from the posterior lip of the cervix. On gross examination mass measured 7x5x4 cms, soft in consistency. The cut surface was gelatinous, slimy, homogenous grayish white with cystic spaces (Figure 1,2). Histopathological examination reveals cervical epithelium which explains the origin of the tumor and a well circumscribed tumor mass in the subepithelial location. The tumor is composed of myxoid matrix, blood vessels, spindle cells. There is abundant myxoid proliferation with stellate and spindle cells. There are occasional cellular areas of



Fig:2

Figure 2: Photograph showing gelatinous cut surface with cystic area.

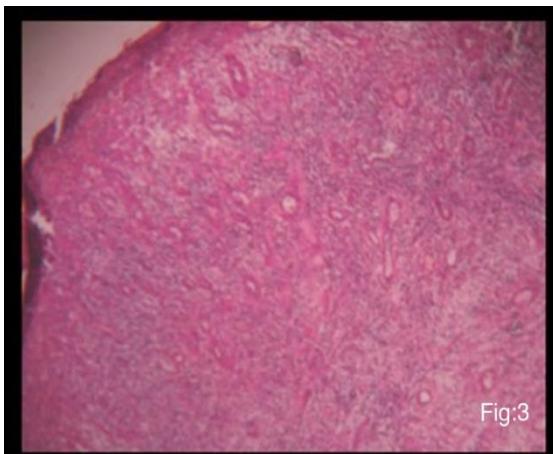


Fig:3

**Figure 3:** Photomicrograph showing subepithelial tumor mass.

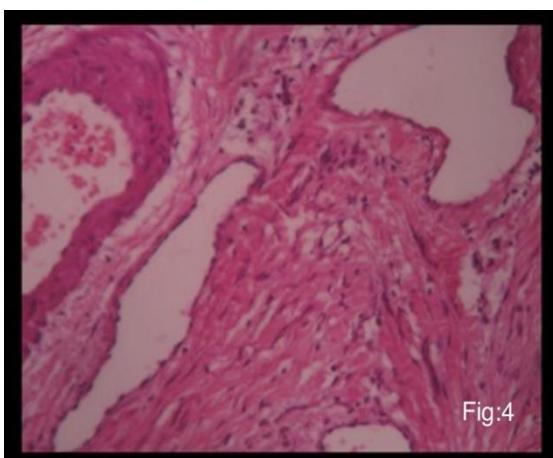


Fig:4

**Figure 4:** photomicrograph showing thin walled blood vessels with spindle cells.

## DISCUSSION

Angiomyxomas are classified either as superficial myxomas or aggressive angiomyxoma.<sup>[3]</sup> Superficial angiomyxomas usually present in the middle age group as a multi lobulated mass in the head and neck region that may be clinically confused with skin tag. The stroma composed of plump spindle and stellate fibroblasts and numerous thin walled blood vessels. On the other hand aggressive angiomyxomas occurs exclusively in the pelvic and perineum region of women of reproductive age group. It is locally aggressive and recurrences after excision is common. Usually tumor is non metastasising , but there are reports of multiple metastasis in whom treated initially by excision and who later succumbed to metastasis disease.<sup>[4,5]</sup> This hormonally responsive tumor is believed to arise from specialized mesenchymal cells of pelvis – perineal region or from multipotent perivascular progenitor cells which often display variable myofibroblastic and fibroblastic features.<sup>[6]</sup> Fibroblastic stromal polyp, superficial angiomyxoma, angiomyxofibroblastoma, cellular

angiofibroma and smooth muscle tumor also need to be considered as a differential diagnosis. Angiomyxofibroblastomas are small well circumscribed submucosal, showing high cellularity. Large amount of thin walled blood vessels and plump stromal cells with very little mucin. Wide surgical excision is the treatment of choice. Recurrence rate varies from 36-70%. Surgery causes significant morbidity due to its frequent occurrence in the lower pelvis and perineum region and its proximity to anorectal and genitourinary structures.<sup>[3]</sup> Recurrence may occur from months to several years after excision (2 months to 15 years).<sup>[7]</sup> Hormonal manipulation with tamoxifen, raloxifene and gonadotropin releasing hormones have been shown to reduce the tumor size and may help to make complete excision feasible in large tumor and in the treatment of recurrences.<sup>[8]</sup> Angiographic embolization may also help in the subsequent resection by shrinking the tumor as well as making it easier to identify it from the surrounding normal tissue.<sup>[9]</sup>

## CONCLUSION

Aggressive angiomyxomas are a rare, slow growing pelvic and perineum mesenchymal neoplasm with marked with marked tendency of local recurrence and but low tendency to metastasize. Wide surgical excision is the treatment of choice but recurrence rate varies from 36-70% with high morbidity rate.

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